Central Deafness Associated with a Midbrain Lesion

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Abstract

Central deafness has been linked historically to bihemispheric involvement of the temporal lobe, with more recent findings suggesting that compromise of other cortical and subcortical structures can also result in this disorder. The present investigation extends our understanding of the potential anatomical correlates to central deafness by demonstrating that bilateral involvement of an auditory structure within the midbrain can additionally result in this condition. Our subject was a 21-year-old male with a subarachnoid bleed affecting both inferior colliculi. Significant auditory deficits were noted for the middle and late auditory evoked potentials, while electrophysiologic measures of the periphery indicated normal function. The patient was enrolled in a rehabilitation program for approximately 14 weeks. Although initially unresponsive to sounds, the patient regained most of his auditory abilities during the 10 months he was followed. This case documents the range of auditory deficits that may be associated with damage to the inferior colliculi, and it profiles a hierarchical recovery of auditory function consistent with test findings.

Key Words: Auditory evoked potentials, auditory processing disorder, central auditory disorders, central deafness, inferior colliculus

Abbreviations: ABR = auditory brainstem response; ALD = assistive listening device; CT = computerized tomography; DL = difference limen; DPOAE = distortion product otoacoustic emissions; MLR = middle latency response; MRI = magnetic resonance imaging

Sumario

La sordera central ha sido históricamente relacionada con el compromiso bihemisférico del lóbulo temporal, aunque hallazgos más recientes sugieren también que el compromiso adicional de otras estructuras corticales y subcorticales pueden producir este trastorno. La presente investigación amplia nuestro conocimiento sobre la correlación anatómica potencial en la sordera central, demostrando que el compromiso bilateral de una estructura auditiva dentro del mesencéfalo puede también resultar en esta condición. Nuestro sujeto fue un hombre de 21 años de edad con un sangrado sub-aracnoideo que afectó ambos colículos inferiores. Una deficiencia auditiva significativa fue notada en los potenciales evocados auditivos tardíos y de latencia media, mientras las mediciones electrofisiológicas periféricas mostraron una función normal. El paciente fue inscrito en un programa de rehabilitación durante 14 semanas. Aunque con una falta inicial de respuesta a sonidos, el paciente

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recuperó la mayor parte de sus capacidades auditivas a lo largo de 10 meses de seguimiento. Este caso documenta el rango de deficiencias auditivas que pueden asociarse con una lesión en los colículos inferiores, y brinda un perfil jerárquico de recuperación de la función auditiva, consistente con los hallazgos en las evaluaciones.

Central deafness is a relatively rare disorder but one that, if appropriately defined and investigated, can add much to our understanding of the specific anatomical structures within the human brain that are involved in the processing of auditory stimuli. Much of what is currently known about the functioning of the central auditory nervous system (CANS) has been gleaned from studies of patients with documented lesions of the brain (Damasio and Damasio, 1989). Investigations that correlate specific auditory deficits with known lesion sites serve to establish clear links between auditory functions and their anatomical correlates. Although significant advances have been made on this front, the exact structures that compromise the CANS in humans have not been completely delineated. This is particularly true of the cortical and subcortical structures that support hearing. Additional studies of patients with CANS damage are needed to build upon the existing knowledge base that details the neural substrate involved in hearing and hearing disorders, and patients with central deafness provide unique opportunities to add to this knowledge base.

Individuals with central deafness often present with a rather dramatic auditory deficit that is not commonly observed in other patients with CANS compromise. Most notably, these patients often demonstrate inconsistent or no responses to sound (Hood et al, 1994; Musiek et al, 1994; Murray and Fields, 2001). Upon initial assessment, these patients may appear to have a peripheral hearing loss since pure-tone testing typically reveals a severe to profound hearing loss—a finding more commonly associated with compromise of the peripheral hearing system. However, if objective test measures of peripheral function are administered, compromise of the periphery is often ruled out, and central auditory system involvement is suggested. With additional testing (e.g., middle and late auditory evoked potentials), the CANS involvement can be documented. Unfortunately, if such testing is not undertaken, then the central origin of the hearing loss is likely to remain unidentified.

To date there have been a number of case studies reported in the literature that have clearly implicated bihemispheric involvement of Heschl's gyrus as the likely origin of the auditory deficits noted in many patients with central deafness (see Musiek and Lee, 1998). In their review of 33 cases of patients with central deafness, Musiek and Lee (1998) found that bihemispheric involvement of the temporal lobe was a common finding in most of these cases. This

observation lends some support to the commonly held notion that damage to the primary auditory areas (i.e., Heschl's gyrus) of both hemispheres must be present in order for the auditory deficits typically associated with this condition to be present. Although bihemispheric temporal lobe compromise was a common finding in the cases reviewed by Musiek and Lee, this site of lesion was not reported for all of the cases reviewed. Thus, the possibility of other lesion sites must be entertained. In addition, their review of the lesion sites for the 33 patients included in their analysis indicated that multiple sites of lesion were involved in nearly every case, with other lesion sites including subcortical structures, the parietal lobe, the frontal lobe, the basal ganglion, the medial geniculate body, and even the pons. With multiple sites of lesions, it is difficult to determine which site, or combination of sites, may have contributed to the central deafness.

One of the challenges in considering the available data on cases with central deafness is lack of consistency in the terminology used. Investigators have used a number of terms to describe their patients, including "central deafness," "cortical deafness," "auditory agnosia," and "word deafness," and in many cases the use of these terms have not been defined fully. For example, "word deafness" has been used to describe a condition wherein patients are unable to recognize words, but the ability to perceive other auditory abilities is not necessarily addressed, leaving the reader uncertain as to the status of this auditory skill in many of the patients studied. On the other hand, "central deafness" has been used to describe patients with a variety of symptoms and test findings, including those that demonstrate a complete lack of response to sounds of any type (including speech), as well as others who show some reductions in hearing sensitivity (hence, not deaf in a strict interpretation), but who show inordinately poor speech recognition abilities or other auditory processing abilities that cannot be accounted for solely on the basis of the noted reduction of hearing sensitivity.

For our purpose we have chosen to use the term "central deafness" to refer to a condition in which the following characteristics are noted: (1) pure-tone thresholds may be absent or they may show severe decrements in hearing sensitivity that do not have an origin in the peripheral hearing system (this may change over time); (2) speech intelligibility is likely to be severely affected by the apparent compromise of hearing sensitivity, and the extent of the speech recognition deficit may be more severe than would have been predicted based upon a consideration of the pure-tone thresholds; (3) objective measures of peripheral and brainstem functions are normal unless the brainstem is involved and contributing to the central deficit; and (4) evoked potentials that assess electrophysiological responses that originate in the cortex or the radiations to the auditory cortex show abnormalities.

As alluded to above, the current lack of specification of the anatomy of central deafness can be attributed to a number of factors. These include the following factors: (1) lesions are seldom specific to structures of interest, (2) radiological evidence in the past has not been exact enough to define the boundaries of damage in auditory system, (3) semantic differences in the terminology used to describe central deafness have precluded precise comparison of the data since it is difficult to define the patient populations in many cases, and (4) the use of limited audiologic testing in many investigations may have resulted in less than a complete elucidation of the nature and the full extent of the auditory deficits in many of the patients studied. Fortunately, recent advances in radiological imaging techniques now permit more careful specification of the location and precise boundaries of lesions within the CANS, and developments in the area of electrophysiologic assessments have contributed to the ability of the audiologist to document auditory deficits arising from cortical and subcortical structures.

A careful review of anatomy of central auditory deafness based upon the available (but qualified) empirical and hypothetical data would argue against the primary auditory areas as being the only auditory regions that could precipitate central auditory deafness (see Musiek and Lee, 1998; Samson et al, 2001). Other cases, with little or no documented involvement of the temporal lobes, have shown that central deafness can result from compromise associated with bilateral infarcts of the insula (Habib et al, 1995), bleeds of the putamen (Nishioka et al, 1993; Makino et al, 1998; Wakabayshi et al, 1999; Taniwaki et al, 2000), third ventricle hydrocephalus affecting the thalamocortical

pathways (Shivashankar et al, 2001), bilateral medial geniculate body involvement associated with a tentorial meningioma (Karibe et al, 2000), arteriovenous malformation embolization involving the inferior colliculi (Vitte et al, 2002), and inferior colliculi destruction and medical geniculate body degeneration associated with a pineal body tumor (Masuda et al, 2000). The present case adds to the growing body of evidence that indicates that compromise of auditory structures outside of the primary cortical areas can result in central deafness.

CASE REPORT

History

This is a report of a 21-year-old college student, who had been in good health with no problems in hearing prior to the time of the medical condition that is described below. He was sent from the college to a local hospital because of severe headaches and vomiting. During his short stay at the local hospital, he became unresponsive and was transferred to a large tertiary care hospital. At the second hospital, he became somewhat more responsive, but he was combative and nonverbal. Key observations at this time included unequal and minimally responsive pupils and purposeful movements to noxious stimuli; however, the patient did not follow commands and was totally unresponsive to verbal stimuli. A computerized tomography (CT) scan at this time showed a subarachnoid bleed. Soon after admission, a diagnosis of Neisseria meningitis was rendered. This rare type of meningitis, to our understanding, can result in accompanying vascular lesions.

During the first two weeks following his medical incident, the patient improved on most fronts. He started vocalizing and saying a few words, which were used appropriately the majority of the time. He was able to move all extremities appropriately and purposefully, and his mental status was improving. He communicated by writing on a pad, which he did reasonably well; however, he remained unresponsive to all sounds. He was soon transferred to a rehabilitation hospital and shortly after the transfer became an outpatient. As an outpatient, he was seen over the next five months for audiological follow-up and auditory therapy as his main

problem over this period of time was his hearing loss.

From a medical standpoint, the patient improved markedly over the first six weeks of his outpatient treatment, and there were no major concerns in this regard. However, he continued to be seen for audiological follow-up following his discharge so that any additional changes in his auditory function could be monitored. His schedule of audiological testing included a number of evaluations that were completed over the first two months immediately following his medical incident, and then subsequent to this he was seen only periodically. The data reported in this study documents the results of most of the formal audiological evaluations that were completed over a 10-month period. Because the patient became fatigued quickly, especially during the initial six weeks, he had to be brought back for repeated visits at various times to complete his audiological evaluations. In addition, the amount of time available for testing was often limited due to a variety of circumstances. Hence, the timelines for testing are not quite as congruent as we would have liked, nor are the evaluations as extensive and complete in all cases as would have been desired.

Magnetic Resonance Imaging (MRI)

At the beginning of his third week in the hospital, an MRI was performed. The MRI showed infarcts at the superior left cerebellum/occipital lobe and the left inferior temporal lobe. Neither of these regions is considered "auditory." However, in the midbrain, both inferior colliculi were almost totally infarcted with some spread of the compromise into the inferior aspect of the superior colliculi (see Figures 1a, 1b, and 1c). A more specific radiologic interpretation indicated that perhaps there was less involvement in the anterior or ventral aspects of the colliculi than in the posterior aspect of the colliculi.

Informal Observations on Hearing

As mentioned earlier, hospital personnel working with this young man noted no response to any sound on the part of this patient for the best part of the first week of his hospitalization. Commencing toward the end of the first week of his hospital stay, and
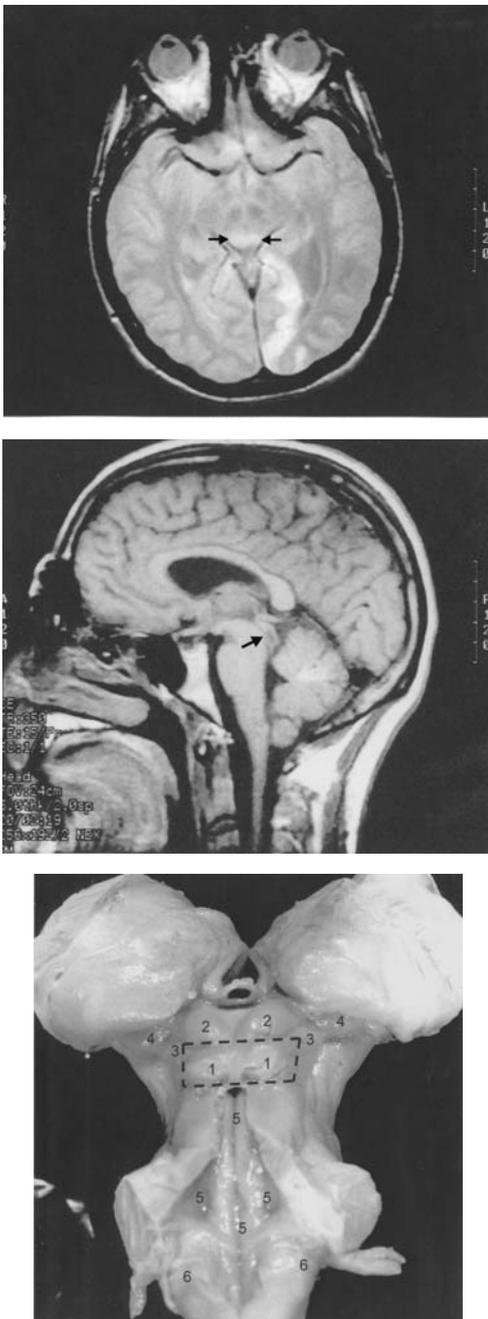
Figure 1. *Top:* a transverse MRI view showing the involvement of both inferior colliculi (arrows), as well as the left lateral aspect of the cerebellum. It appears that there may be slightly greater area involvement for the right inferior colliculus compared to the left. *Middle:* a mid saggital MRI showing the involvement of the inferior colliculus (arrow). *Bottom:* a human brain specimen that emphasizes a posterior view of the pons, midbrain, and thalamus. The area inside the dashed box reflects the authors' interpretation of the involved anatomy in this study's subject. The region of involvement encompasses both inferior colliculi and the inferior fringe of the superior colliculi. Key: 1 = inferior colliculus, 2 = superior colliculus, 3 = brachium of the inferior colliculus, 4 = medial geniculate bodies, 5 = fourth ventricle, 6 = cochlear nucleus area. Courtesy of W. T. Mosenthal and F. E. Musiek.

continuing for several weeks thereafter, the patient claimed he heard a certain sound, yet other people in the room with him did not hear the sound. Beginning in his second week, the patient acknowledged hearing some sounds in his environment that were actually present, but he could not identify the exact nature and/or the source of these sounds. It appeared that the sounds that were heard were relatively loud and of a broad spectrum. By the third week of his hospital stay, definite improvements in hearing were noted. The patient could hear voices, but he could not understand anything that was being said to him. He stated that "voices don't [didn't] sound like voices." He was speaking often, but hospital personnel noticed that his voice was monotone. Listening was a strain, and it fatigued him to the point where he often complained about it. He continued to hear environmental sounds, such as a dish being dropped and water running from a faucet, and he seemed to be able to discern loud sounds from soft sounds. However, he continued to be inconsistent in his identification of the source(s) of environmental sounds. The patient additionally complained of extreme difficulty listening to people speak whenever background noises were present. At this point in his recovery, he was able to read slowly, and this skill has continued to improve over time.

In the fourth week post–medical incident, the patient reported that he felt his hearing was improving. He could hear voices on TV but could not understand what was being said. In general, the patient knew when people were speaking but could not follow speech on a consistent basis. He stated that riding in a car was a difficult situation because everything was too loud. He was identifying more environmental sounds and was encouraged when he identified the clicking of high heels caused by someone walking on a hard floor. There remained some disassociation between what the patient believed he heard and what sounds were actually present in his environment. He claimed to be able to hear women's voices better than men's, and he reported that he could hear parts of his girlfriend's voice on the phone. During this time period, the patient began to experience bilateral tinnitus (ringing type sound), which he reported was worse in the morning than later in the day.

In the time period from about 5 through 12 weeks, the patient's hearing continued to

show steady improvement. He could now understand several words, and this ability gradually improved over time but was highly dependent on the patient's being located in a quiet environment. Background noise, even if of a low intensity (e.g., noise generated by a small fan), caused considerable problems in his understanding of speech. He noted that he heard better when he was actively participating in a conversation compared to when he was listening passively. He reported that speech seemed muffled and described his listening experiences as if he were listening to speech over a two-way radio transmitter. He could hear birds chirp but described the chirping of the birds as having a "robotic" quality. His therapist noted that his voice was softer at this time, and possibly less monotone.

In the post–hospital admission period from 12 weeks to 10 months, the patient was seen only a few times formally. His ability to understand speech in quiet continued to improve, and he was able to follow most conversations. Complex and similar sounding words remained difficult for him to correctly interpret and discriminate, and background noise continued to be a major detriment to his accurate understanding of speech. The patient's confidence in communication improved considerably over this period of time, and at this point in his recovery he had gone back to college and appeared to be doing satisfactorily with some accommodations such as preferential seating. As part of his rehabilitation program, the patient had been provided an assistive listening device (ALD), which provided some assistance; however, there was some reluctance on the part of the patient to use it consistently as he did not want to become dependent upon the device. The patient was motivated to act as if he has no problem at all and therefore downplayed use of the ALD, as well as some of the academic accommodations that were recommended. His auditory behavior at 10 months after his medical incident was similar to that of an individual with a moderately severe high-frequency hearing loss. However, background noise continued to be a major problem, more so than would be expected for a person with only a moderately severe high-frequency sensorineural hearing loss.

PROCEDURES

Pure-Tone and Speech Audiometry

All audiological tests were conducted in sound-treated rooms. Conventional pure-tone air- and bone-conduction threshold techniques were used to establish hearing thresholds for this patient; however, at times these had to be modified to accommodate the patient's needs. Spondee thresholds were obtained on some occasions using only a small subset of spondees as indicated in the test results section. Speech recognition measures were conducted using the Northwestern University Test Number Six (NU-6). Speech audiometry was carried out using monitored live voice procedures as this mode of presentation seemed most clinically feasible particularly during the early evaluations of the patient's hearing.

Immittance Audiometry

Tympanograms and acoustic reflexes were derived in a conventional manner using a GSI 1733 middle-ear analyzer. Acoustic reflexes were measured both ipsilaterally and contralaterally at 500, 1000, and 2000 Hz.

Otoacoustic Emissions

Distortion product otoacoustic emissions (DPOAEs) were derived for this patient. Levels (L1, L2) were 65 dB SPL and 55 dB SPL, and the f1 to f2 ratio was 1.2. The left ear DPOAEs were measured for frequencies from 600 to 8000 Hz, and the right ear for frequencies from 800 to 8000 Hz (the lower frequencies could not be measured in the right ear due to the presence of excessive noise at these frequencies). Four distortion products per octave were obtained for each ear (see Smurzynski, 1994 for details of this procedure).

Auditory Brainstem Response (ABR)

All ABR testing was conducted using 100 μsec clicks presented through TDH-39 earphones to each ear independently at 85 dB nHL at a rate of 15.7 clicks per second unless otherwise noted. Neuroelectrical activity was recorded from electrodes attached to the high forehead and each earlobe (one reference and

one ground) and averaged over 2000 trials. Impedance across the electrodes was maintained at less than 5 kohm. Filter bands for the ABR were set at 150 to 3000 Hz with a 12 dB per octave roll-off. Responses were analyzed over a 10 msec time period.

Middle Latency Evoked Response (MLR)

The MLR was obtained using 100 µsec clicks presented through TDH-39 earphones at 70 dB nHL at a rate of 8.7 clicks per second for each ear. For the initial test session, electrodes were placed at Cz and referred to either A1 and A2; however, all subsequent MLR recordings were conducted with electrodes at Cz, C5, and C6 with the same references. Impedance across the electrode array was maintained at less than 5 kohm. In all cases the reference electrode was on the side of the head that was receiving the click stimulus. There were 800 accepted trials conducted for each run, and filtering was 20 to 1500 Hz with a 12 dB per octave roll-off. The time period for analysis was 70 msec.

P300 Potential

The P300 was conducted in each ear using an odd-ball paradigm with 1000 Hz and 2000 Hz tones representing the frequent and rare stimuli, respectively. These tones had a 10 msec rise and fall time and a 20 msec plateau and were presented at 70 dB nHL through TDH-39 earphones at a rate of 1.2 stimuli per second. Electrical activity from the scalp was picked up by electrodes placed at Fz, Cz, and Pz. Stimuli presentation rates were set at 80% for the frequent stimulus and 20% for the rare stimulus, and the total number of stimuli to be accepted during each run was fixed at 300. Impedance across electrodes was less than 5 kohm. Filtering was set at 1 to 30 Hz with a 12 dB per octave roll-off. The time window was 800 msec. The patient had to count the number of rare stimuli and report this number at the end of each run (a score of >80% correct behavioral counting performance was required for the acceptance of an electrophysiological response). Responses from the rare and frequent stimuli were averaged separately. The frequent stimuli yielded late potentials N1 and P2, and the rare stimuli provided N1, P2, and P300 waves for analysis.

Intensity Discrimination

Intensity discrimination testing was conducted in each ear for three frequencies (500, 1000, and 2000 Hz). Each test stimuli consisted of the presentation of two tones that were delivered at either the same intensity level or at different intensity levels, and the patient was asked to indicate if the tones were the same or different in loudness. Each tone was 500 msec tones in duration with a 40 msec rise-fall time, and the interstimulus interval between the two successive tones was approximately 1 sec in duration. Trials of equal versus different intensities were presented at random. The reference presentation level was fixed at 50 dB HL; thus, in each test stimuli, one of the tones was presented at this intensity level while the second tone was varied for the purpose of identifying the smallest intensity difference that the patient could recognize. The patient was required to achieve two out of three correct judgments at a particular intensity difference before going further. This procedure was repeated several times, and the average of difference limens (DLs) obtained was computed. At times this procedure had to be modified to accommodate the patient. For example, on some days, more practice items with large DLs were provided and/or the presentation approach was slightly changed to allow a better orientation toward the test.

TEST RESULTS

Basic Audiological Evaluation

The first formal audiological evaluation was performed about two and one-half weeks after admission. At this point, the patient's hearing had already improved considerably from the first week; however, in spite of this noted improvement in hearing, pure-tone thresholds were difficult to obtain, especially at the low frequencies with a frequent occurrence of false positives. This difficulty with the assessment of pure-tone sensitivity continued for most of the early evaluations, and to a lesser degree for later evaluations. One of the factors that contributed to the difficulty with determining thresholds for pure tones was that the patient noted little or no tonality in the stimuli; hence, the sounds
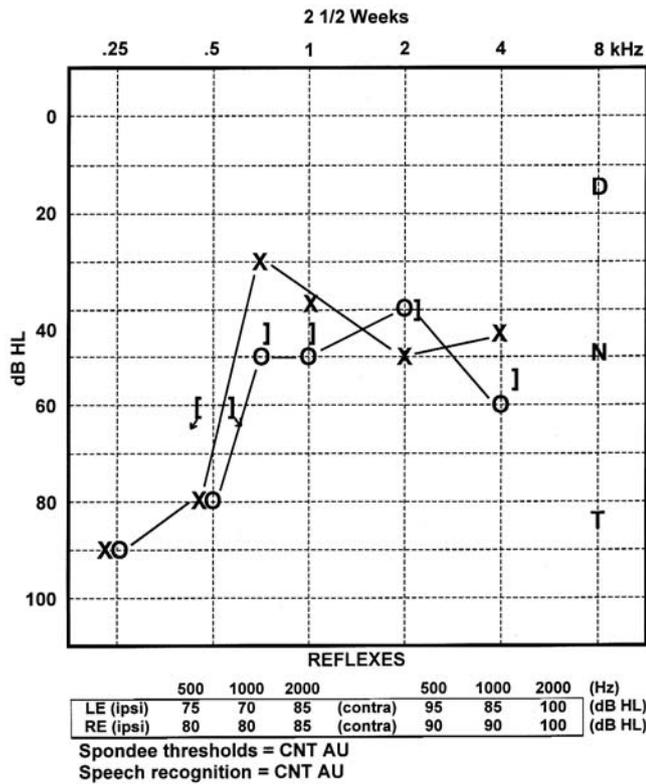
Figure 2. The initial audiogram and acoustic reflex test results for a 21-year-old male with bilateral involvement of the inferior colliculi. Spondee thresholds and speech recognition could not be tested (CNT) for either ear (AU) at this time. Key: X, O = left and right ears, [ ] = bone conduction right and left sides, DNT = did not test, AU = both left and right ears, CNT = could not test, bone conduction was not tested at 250 Hz.

Figure 3. An audiogram showing pure-tone thresholds, spondee thresholds for selected spondees, speech recognition scores, and acoustic reflex findings. These results were obtained at approximately three and one-half weeks following the patient's hospitalization.

were not distinctive. This was particularly problematic during the first few evaluations, especially for low frequencies and the right ear. Another factor that was likely to have affected his performance during this initial evaluation, as well as in other subsequent evaluations, was the difficulty that the patient reported in maintaining his attention. At the time of his initial evaluation, a severe low-frequency sensorineural loss, rising to a mild to moderate loss, was indicated bilaterally (Figure 2). Speech audiometry could not be obtained during this evaluation for either ear, but tympanograms were of normal pressure, compliance, and shape bilaterally. Ipsilateral and contralateral acoustic reflex thresholds were grossly within the normal range for absolute threshold levels for both ears, but these thresholds were at reduced sensation levels in reference to pure-tone thresholds for both ears at 500, 1000, and 2000 Hz.
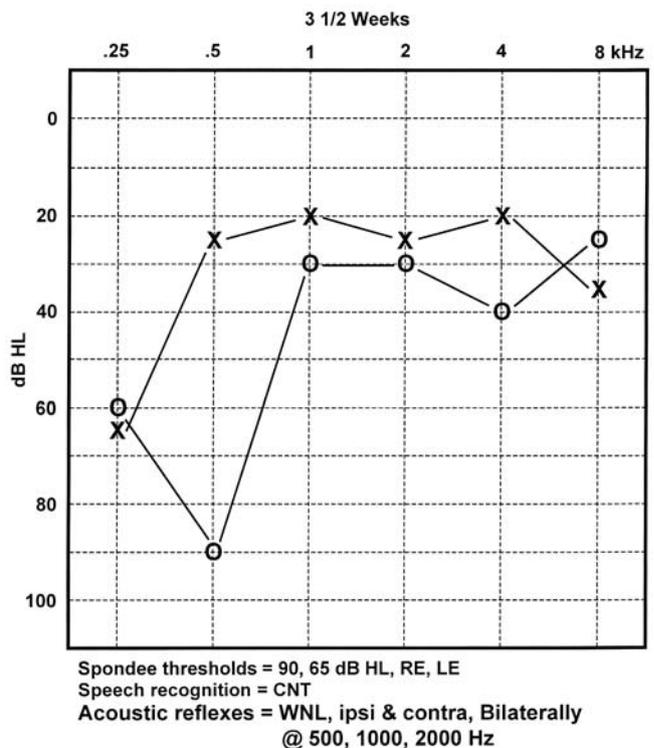
The next formal evaluation was performed at three and one-half weeks. As can be seen in Figure 3, there was a definite improvement in the hearing thresholds for both ears. Spondee thresholds were obtained bilaterally for the first time using selected spondees; however, these thresholds were elevated and in poor agreement with the pure-tone averages for both ears. Speech recognition could not be tested at this time as the patient could not understand even the carrier phrase. Acoustic reflex and tympanometric test results remained normal bilaterally.

The next audiological assessment was conducted at five weeks (Figure 4), and although it showed improvement at the mid and high frequencies, the low frequencies were difficult to assess. There was so much variability in performance at the low frequencies for the right ear that thresholds could not be established. The patient claimed that the low-frequency tones (especially for the right ear) "sounded like the wind blowing tall grass in a field." Speech recognition could
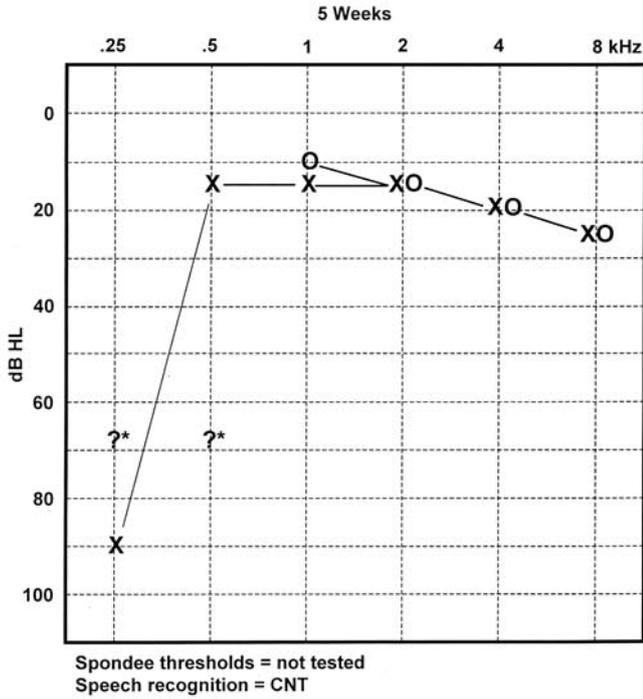
5 Weeks

Figure 4. An audiogram and speech recognition results at five weeks. The "?*" indicates the "best guess" thresholds for the right ear at these frequencies as threshold criteria could not be achieved at this time due to variability in responses and fatigue on the part of the patient.
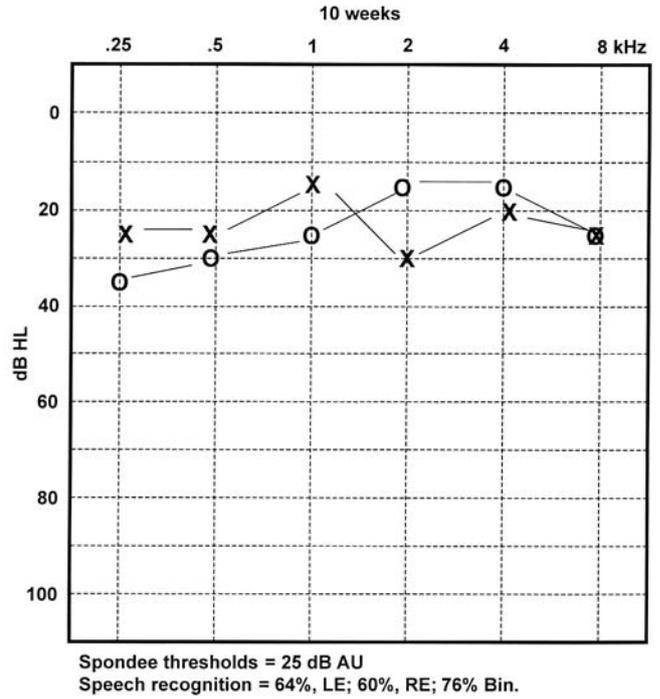
10 weeks

Figure 5. An audiogram, spondee thresholds, and speech recognition scores for each ear and in the binaural (Bin.) condition obtained at 10 weeks.
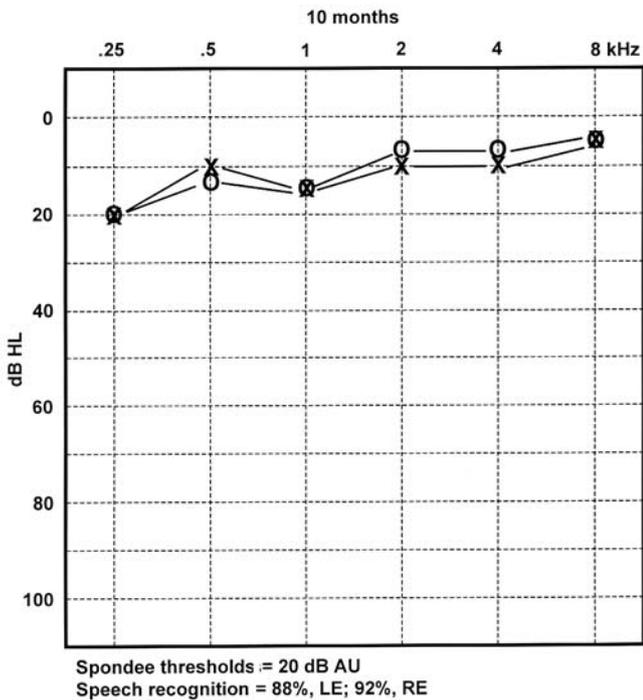
10 months

Figure 6. An audiogram at 10 months with spondee thresholds and speech recognition scores.

not be tested. The patient became very fatigued during this evaluation, and his motivation was dwindling.

The next audiogram was at 10 weeks (Figure 5). Pure-tone thresholds had improved to close to normal bilaterally at this time and were in good agreement with the spondee thresholds bilaterally. At this evaluation, speech recognition could be tested for both ears and in a binaural (diotic) condition for which the best score was obtained. At approximately five months, speech recognition had improved into the 80% range bilaterally as informal assessments during his rehabilitation period were conducted. He was discharged from therapy at five months. At 10 months he was seen for assessment and showed essentially a normal audiogram and speech testing (Figure 6). As mentioned earlier, however, he continued to experience difficulty with background noise.

Otoacoustic Emissions

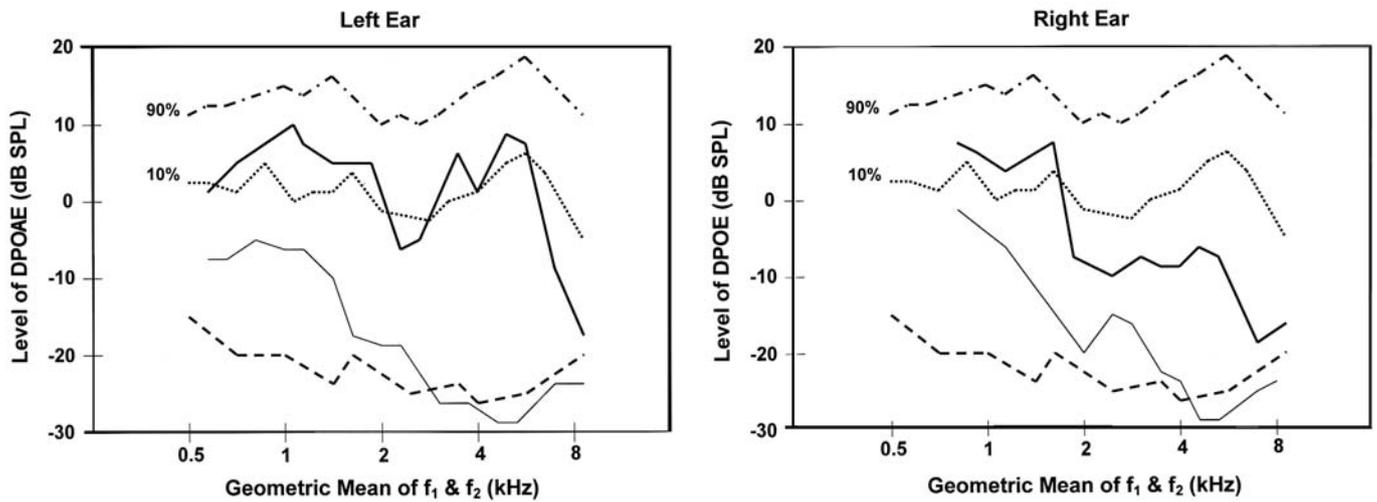Distortion product otoacoustic emissions were administered approximately four weeks

Figure 7. DPOAEs responses for the left and right ears. The heavy solid line is the patient's DPOAE levels; the thin solid line is the patient's noise floor; the dashed line is the average noise floor; and the dotted line and dashed/dotted lines are 10th and 90th percentiles.

after hospital admission. The left ear results were essentially normal for absolute DPOAE levels and DPOAE to noise floor measurements for frequencies 600 through 6000 Hz (Figure 7), whereas the right ear results were at normal absolute levels from 800 through 2000 Hz, but at abnormal levels for the higher frequencies tested (Figure 7). The right ear DPOAE to noise floor measurements were grossly normal for all test frequencies (800 through 6000 Hz). The patient was anxious and tense during this evaluation, and reliable measurements could not be made below 800 Hz due to excess noise.

Auditory Brainstem Response (ABR)

The initial ABR was performed at three and one-half weeks and showed normal waves I, II, and III bilaterally. It appeared that the IV-V complex was compromised for both ears. It is possible that wave IV could be normal, but the amplitude and morphology of V is not as would be expected for either ear (Musiek and Lee, 1995). The amplitude of the IV-V complex is less than half the amplitude of wave I bilaterally. This abnormal V-I amplitude ratio highlights the compromise of the IV-V complex (Musiek and Lee, 1995). The ABR was evaluated several times over the recovery period up to 10 months but remained essentially the same as shown in Figure 8.

Middle Latency Response (MLR)

The initial MLR was conducted at three and one-half weeks and showed no readable response for either ear (Figure 9). A second MLR derived one week later again revealed no repeatable responses for either ear. The first follow-up MLR that showed a notable change was derived at 24 weeks (Figure 10). At this time, a meager Pa wave began to emerge at essentially normal latencies for both ears across all electrode sites. It was difficult to determine if an Na wave was present at this time, but it seems doubtful.
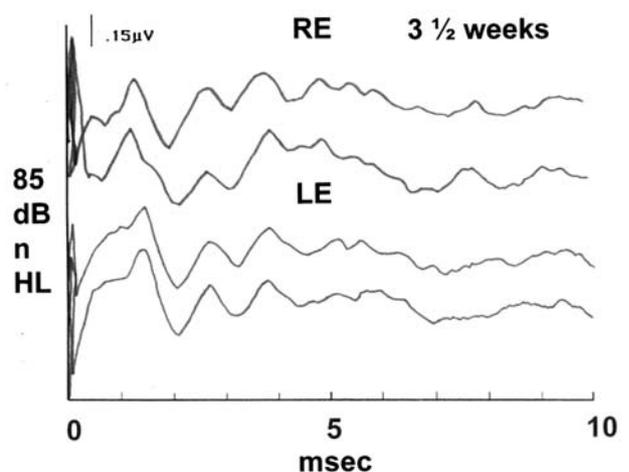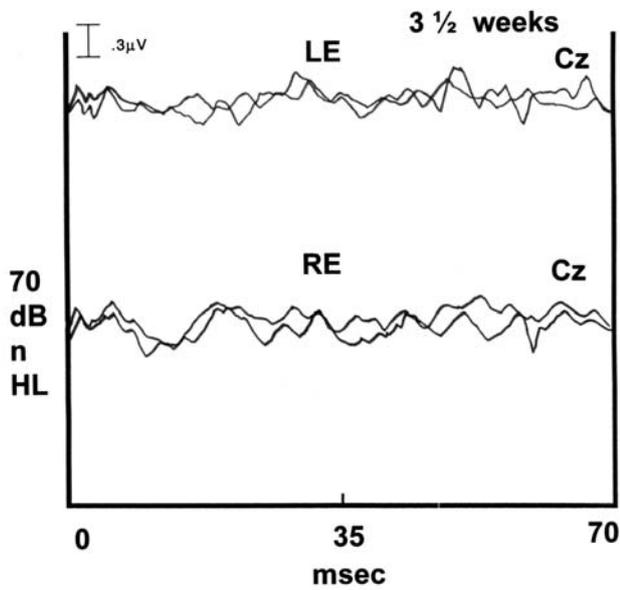

Figure 8. The initial ABR.
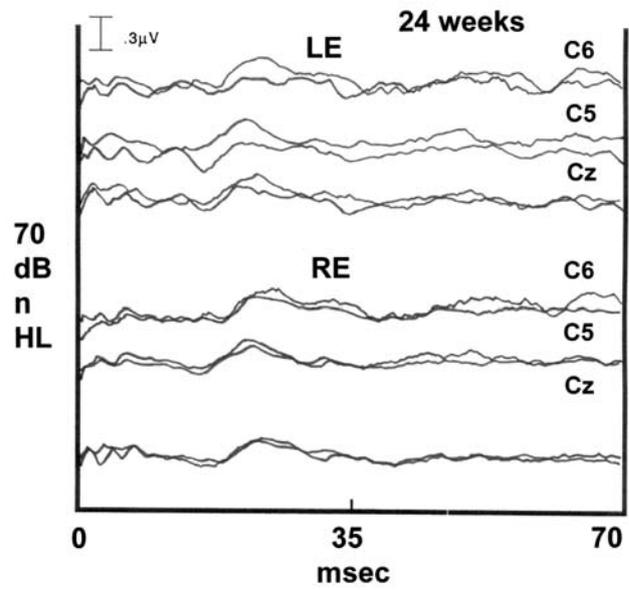
Figure 9. The initial MLR.

Figure 10. An MLR obtained at 24 weeks.

The Nb and Pb waves were absent. At 10 months, the MLR was more noisy but showed similar responses to those noted at 24 weeks.

P300 Potentials

The P300 as well as the N1 and P2 late potentials (as derived using an odd-ball

Figure 11. The initial findings for the P300 as well as late auditory evoked potentials (N1 and P2).

paradigm) showed no readable responses at four weeks (Figure 11). An evaluation at 9 weeks showed a P300 emerging for both ears across all electrode sites (Figure 12). The P300 was within the normal latency range for the left ear stimulation but was extended for the right ear stimulation. The P2 also began to emerge, though attenuated, at Fz and Cz electrode sites for both ears. The latency of P2 was extended (240 msec), and an N1 was not seen for any recording site at nine weeks. At 10 months, the P300 increased in amplitude and decreased slightly in latency across the Pz and Cz electrode sites for both ears. The N1 and P2 for both ears remained essentially the same for Fz and Cz electrode sites; however, at 10 months a P2 emerged for the Pz electrode position (Figure 13).
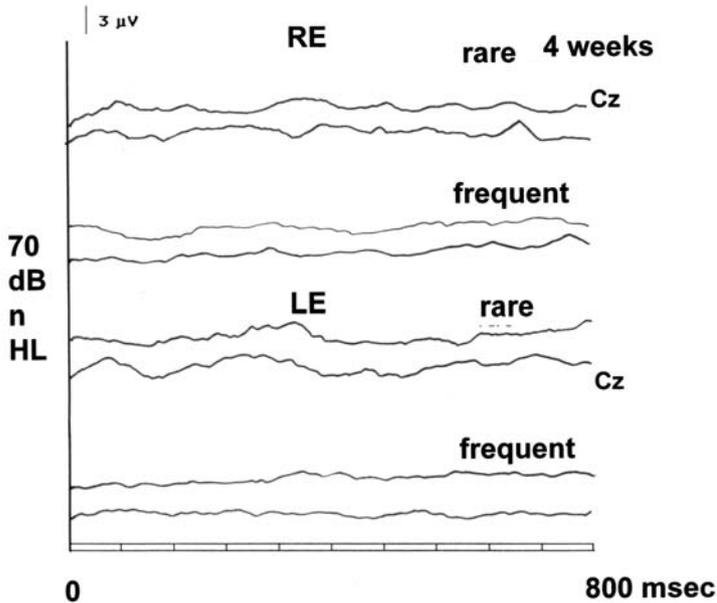
Intensity Discrimination

Intensity discrimination was measured at 5 and 11 weeks. As depicted in Figure 14, there was considerable improvement in intensity discrimination over this period of time. The right ear, which was the ear with poorer performance at five weeks, improved more than the left ear. At five weeks, the right ear could not be assessed due to too great of a hearing loss. The patient performed better in the binaural condition for both test sessions (Figure 14). The cut-off for normal
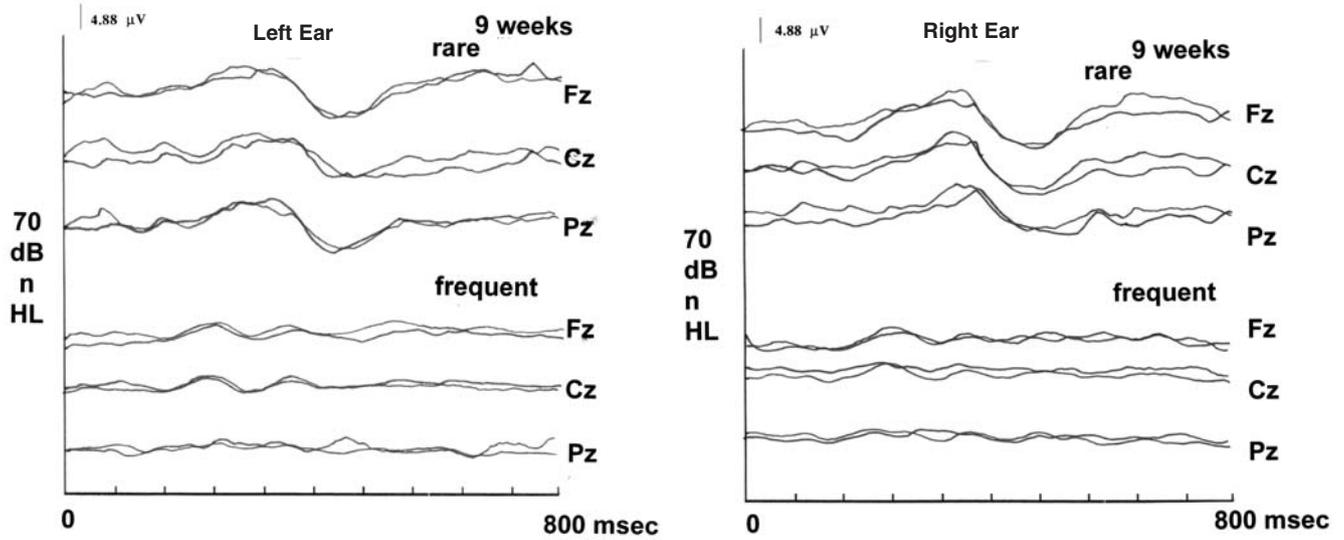
Figure 12. Left and right ear results for the P300 and late auditory evoked potentials at nine weeks.

performance on this task is 2 dB for both monaural and binaural presentations.

REHABILITION PROGRAM

The aural rehabilitation program commenced approximately one month after hospital admission and continued on a consistent basis for about four and one-half months. Individual therapy sessions were held once per week for one to one and one-quarter hours, and the patient's performance on the various therapy tasks outlined below was monitored throughout this period. Task difficulty was governed to be in the moderate range so that overall the tasks were not too demanding or too easy and the patient was either directly or indirectly provided feedback as to his performance during training procedures. The patient was also given therapy assignments to complete at home. These included such activities as auditory directives, discrimination tasks, and critically listening to music. The description of the formal therapy has been divided into
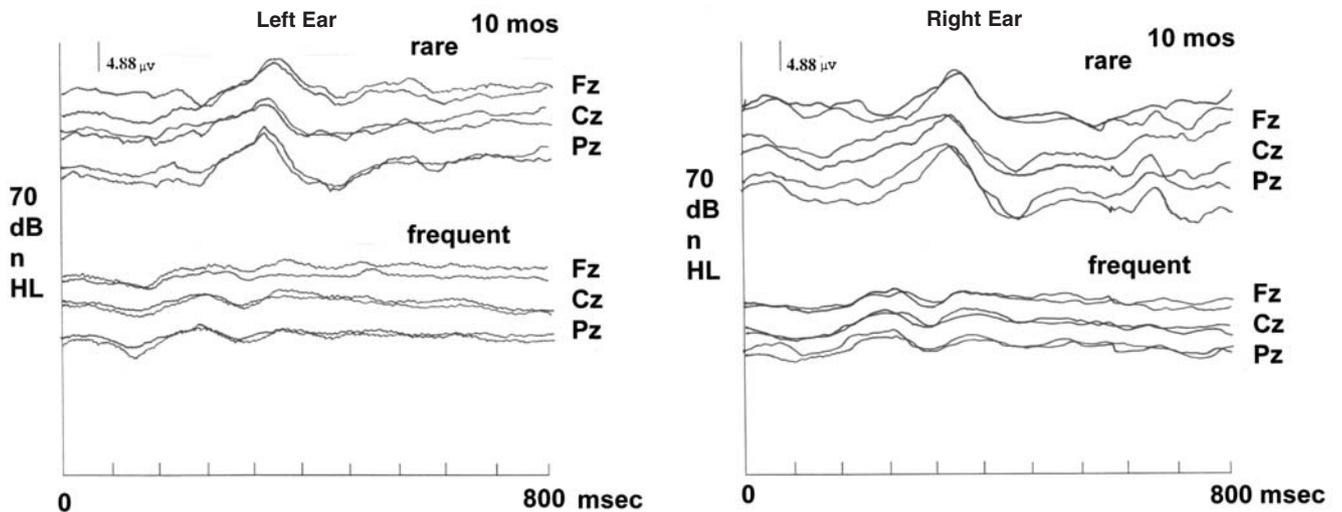

Figure 13. Left and right ear results for the P300 and late auditory evoked potentials at 10 months.
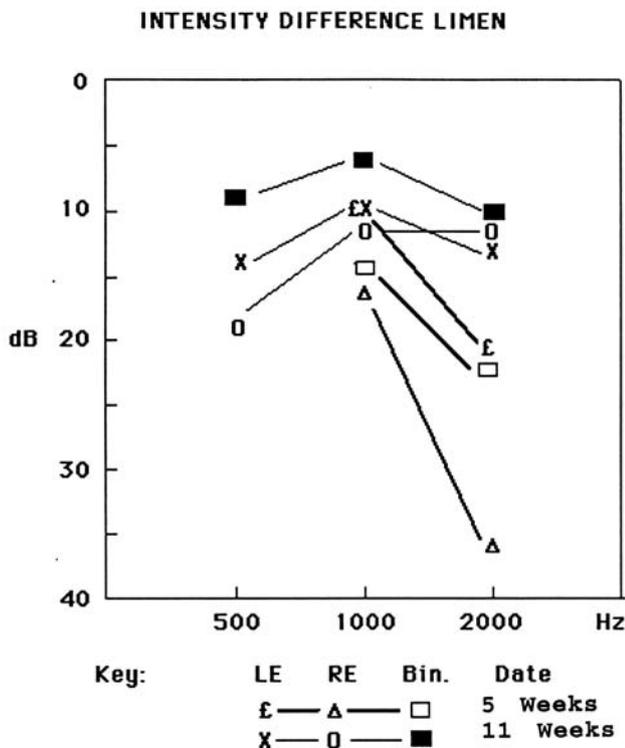
Figure 14. The intensity DL derived at 500, 1000, and 2000 Hz in each ear at 5 weeks and at 11 weeks. At 5 weeks the DL at 500 Hz could not be recorded for the right ear.

three time periods; early (approximately the first month), mid (approximately the next two months), and late (approximately the last month and a half).

Early Therapy Sessions (with Visual Cues)

One of the first therapy goals was to work on the patient's vocal intonation as the patient's voice was monotone. Early therapy activities additionally focused on having the patient respond appropriately to fundamental questions about himself. Therapy on identifying and discriminating speech and environmental sounds was a major component of this therapy period. The patient was asked to make judgments as to intensity differences between and/or among sounds both in formal and informal paradigms. In addition there was some work on speech reading and emphasizing the use of visual cues. Much therapy was centered around multimodality stimulation and associating cues across different modalities.

Midtherapy Sessions (with and without Visual Cues)

In this therapy period, the patient was asked to identify familiar voices and to try to discriminate male from female voices. Work was continued on identifying and discriminating environmental sounds that, for the most part, were tape recordings that provided a wide variety of acoustic stimuli. Since the patient liked popular music, some therapy procedures centered around identifying songs and their artists. The patient also spent time repeating sentences that were presented to him in the presence of background noise that was varied in terms of noise type and level. Toward the end of this therapy period, auditory training consisted of identifying vowels, blends, and consonants using Sloan's (1986) materials. Discriminating homophonous words and contrasting nonsense words with real words in both monaural and binaural listening conditions was also conducted. The patient was asked to read aloud and listen to himself, and intensity discrimination tasks were continued. Finally, a portion of the therapy time was often devoted to work on one-, two-, and three-step auditory directives.

Late Therapy Sessions (with and without Visual Cues)

At this point, the patient had made considerable improvement, and, accordingly, the tasks were made more challenging. Discrimination tasks with similar sounding phonemes, consonant-vowels (CVs), vowel-consonants (VCs), and homophonous words became a major focus in therapy. Identification tasks involving the presentation of words and sentences in noise were continued, and the patient was also asked to identify and correct words that were purposely distorted in a sentence. The patient claimed he had difficulty in group discussions recognizing when he was being spoken to and responding appropriately. He was provided training on using visual cues in these situations to help him determine when he was expected to be part of the conversation. An assistive listening device (ALD) was introduced, and although the patient experienced some success with this device, he was not in favor of its long-term use. Shortly after the formal therapy sessions ended, the

patient went back to college with an ALD. He was also provided a note taker for his classes.

DISCUSSION

Cases of central deafness are indeed a rarity. Usually most cases of central deafness involve vascular damage to the primary auditory area in both cortices (Musiek and Lee, 1998). Even more unusual is central deafness associated with lesions of the brainstem or subcortex. Unlike the majority of cases of central deafness that result from bilateral auditory cortex lesions, this case had healthy (auditory) cortical and subcortical substrate. In this case, the auditory deficits noted were most likely related to impulse disruption arising at the midbrain level, which deprived the cortex of appropriate neural input from both sides of the midbrain. This case is one that showed recovery of many, but not all, higher-level hearing processes. By following this patient for 10 months, information as to the nature of the recovery process was gained.

Informal and Formal Observations of Hearing: Course of Recovery

We believe there is a strong possibility that this patient may have been totally centrally deaf during the first week to week and a half post–admission to the hospital. Hospital personnel reported that the patient was unresponsive to acoustic stimuli during this period of time. By the time the first hearing test was completed at two and one-half weeks, the patient had recovered considerable hearing ability, but he remained essentially word deaf throughout the first month following his medical incident. Suffice it to say, the involvement of the inferior colliculi created a major deficit across all aspects of hearing.

The informal observations of this patient's hearing recovery reflected a logical progression, which began with the return of the ability to perceive simple sounds and increased to the perception of more complex sounds (speech). Initially the patient could not detect any sounds; however, within the first week he could detect sounds but could not identify them. The detection and subsequent identification of simple environmental sounds was followed by the recognition of more complex and less familiar acoustic stimuli. By

10 weeks post–hospital admission, the patient could communicate reasonably well when visual cues were optimized, and the acoustic environment was ideal. The patient's improvement for hearing in poor acoustic (noisy) environments was minimal in the first few weeks after his medical incident, and even at 10 months this continued to be a major problem for this patient.

Formal audiological testing showed a similar progression of recovery. The simplest stimuli, pure tones, were the first to show progress in the recovery process and ended up in the normal range bilaterally. The spondee thresholds eventually progressed to within the normal range, as did speech recognition ability although the latter did require more time. The evoked potentials, other than the ABR, showed improvement over time. However, none of the evoked potentials recovered to normal status. This progression of regaining auditory perception for simple and then more complex processes is consistent with what has been reported previously in the literature for patients recovering from CANS damage (Mendez and Geehan, 1988; Nishioka et al, 1993; Musiek et al, 1994). The more complex the acoustic stimulus, the more neural substrate is needed. This has been well demonstrated in dichotic listening paradigms in people with temporal lobe damage (Kimura, 1961). In cases of central deafness, neural damage is extensive, and recovery of sufficient neural substrate to support complex processing requires an extended period of time (Mendez and Geehan, 1988).

This case opposes the classic concept that central auditory lesions do not affect pure-tone thresholds. This case and other studies on both humans and animals demonstrate that central auditory involvement can result in the reduction of pure-tone sensitivity (Heffner and Heffner, 1986; Musiek et al, 1994; Murray and Fields, 2001). Whether or not pure-tone sensitivity is affected by central auditory involvement depends on the site and size of the lesion and how soon the hearing sensitivity is measured after the damage occurred (see Heffner and Heffner, 1986).

An interesting observation made on this patient, especially early on in the recovery process, was that he would think that he heard sounds that really did not exist. This behavior has been seen in centrally deaf

patients before (Musiek et al, 1994). We cannot be sure if this phenomenon occurs because the sounds are imagined in some manner, such as an auditory illusion or hallucination, or whether they are the result of some other psychophysiological phenomenon. Nonetheless, the patient genuinely believed the sound(s) was indeed present in his environment. The incidence of this misinterpretation or illusion of sound appeared to decrease as the patient's hearing improved.

Anatomy and the MRI

This case is one that has some rather specific neuroanatomy. Although there were lesions in other areas of the brain, the only significant auditory involvement was at the right and left inferior colliculi. There may have been some "minor" involvement in the more caudal aspects of the superior colliculi, which may have some auditory function. It has been demonstrated that in some animal species the superior colliculus may have some auditory fibers. These, however, are in the minority (Irvine, 1986). Therefore, the lesion site in this case is one that is quite specific, which helps in determining the role of the inferior colliculus on the various aspects of hearing function studied in the present investigation.

Close inspection of the MRIs may indicate that there was less damage at the more anterior aspect of the colliculi. If this was in fact the case, it may explain why there was better hearing at the high frequencies than at the low frequencies. The tonotopic arrangement of the inferior colliculus has been shown to progress from high to low frequencies in a posterior to anterior manner, with high frequencies located deep in the midbrain (Merzenich and Reid, 1974). If the deep substrate was damaged less, it may have allowed the coding and progression of the high-frequency stimuli up the remainder of the auditory pathway. The severe damage to the low-frequency region of the inferior colliculus may have prohibited frequency coding at this level and may have also either stopped or disrupted the rostral flow of low-frequency impulses to the thalamo-cortical tract. This may also explain why the patient reported during testing that the low-frequency pure tones did not sound tonal.

Auditory Evoked Potentials

Major damage to the inferior colliculus poses a situation that is of interest to the ABR, MLR, and late potentials. It was once thought that the inferior colliculus was the major generator of the ABR wave V. Studies on humans during neurosurgery changed this thinking (Moller, 1983; Moller et al, 1995). Current thinking is that wave V may have a slight contribution from the inferior colliculus, but that it is primarily generated by the lateral lemniscus (see Hall, 1992). This case adds information to what is currently known about the wave V generator site. The radiology shows that the damage was limited to the inferior colliculus and possibly the inferior portion of the superior colliculus but that the pons was spared in this patient. Although wave V is present, it is clear that the waveform morphology is somewhat compromised. This finding was repeated a number of times. Therefore, it appears that in this patient wave V may have some contribution from the inferior colliculus in addition to contributions from neural structures located more caudally. The ABR IV-V complex is clearly distorted in spite of the fact that wave IV can be discerned from wave V. In addition, the IV-V complex is significantly smaller in amplitude than wave I in both ears. This finding has often been considered an indicator of a brainstem or retrocochlear lesion (Musiek and Lee, 1995). The early waves of the ABR are normal bilaterally indicating good neural integrity in the region of the caudal pons.

It is interesting to note that the ABR waveform did not change over time and hence did not reflect the improvement noted in the patient's behavioral hearing. This disassociation may be related to several factors. The auditory fibers in this area (midbrain) may have recovered to some degree and rerouted themselves around the damaged area, or new fibers may have been recruited to assume some of the functions of the damaged neurons. It has been shown that the inferior colliculus has the ability to reorganize rather quickly when deprived of peripheral input (Wang et al, 1996). In this patient, this new or recovered neural substrate may have supported the processes necessary for the improvements noted in his auditory behaviors but not have achieved the neural synchrony or dipole orientation

necessary to generate a normal ABR wave V. We also know there is not a direct relationship between behavioral hearing and the ABR (Worthington and Peters, 1980). Hence, behavioral changes may be reflected in different measures than the ABR. The ABR is dependent on neural synchrony, and perhaps the fibers responsible for this synchrony did not change but, rather, the change may have occurred in neural substrate not involved directly in the generation of the ABR.

The MLR that was recorded at three and one-half weeks showed no readable response for either ear at the Cz recording site. At 24 weeks there clearly was some recovery of the MLR with the Pa wave visibly discernable for both ears and across three electrode sites (Cz, C5, C6). The tracings from the right ear appeared to be slightly more synchronous than those derived from left ear stimulation. Although there was some definite recovery at 24 weeks in the MLR waveform morphology, it did not continue to improve noticeably beyond this time as waveforms at 10 months were similar to those noted at 24 weeks. The improvement in the MLR waves would again support the notion that synchronous impulses began making their way to the thalamo-cortical regions of the brain (see Kraus et al, 1994). In the initial MLR, this may not have been the case. Again, the neural pathways in the midbrain may have recovered or been reorganized to allow synchronous impulses to reach the generators of the MLR. Without some form of recovery or auditory plasticity occurring, it appears that bilateral damage to the inferior colliculus may preclude recording an MLR. Besides the auditory thalamo-cortical pathways, the MLR waveform may be dependent on the reticular formation (see Kraus et al, 1994). Therefore, it may be possible that compromise or a "shutting down" of the reticular formation may have compromised the MLR. There was no asymmetry for the MLR waveforms across electrode sites, indicating that the "effect" was bilateral. This is consistent with the lesion site being bilateral and symmetrical. It is also interesting to note that the MLR improved without any change in the ABR late waves (IV-V). Therefore, one might conclude that the MLR change was not dependent on the same subcortical/brainstem substrate as the ABR at least in this case. In this regard, it has been shown previously

that an MLR can be present with an absent ABR (Squires and Hecox, 1983).

The P300 as well as the N1 and P2 late potentials were absent at the Cz recording site at the initial evaluation, and there was essentially no difference between the frequent and rare waveforms. However, at nine weeks the P300 did emerge along with rather meager N1 and P2 complexes across all electrode sites for both ears. The late potentials all appeared to be delayed in absolute latency. As with the MLR, these late evoked potentials may have initially been absent due to the lack of synchronous input secondary to severe bilateral CANS damage at the midbrain level. The emergence of the N1, P2, and P300 potentials at nine weeks may indicate (as with the MLR) the reestablishment of a functional neural pathway in the midbrain that could provide synchronous input to the subcortex and cortex. It seems important to point out that without this "reestablished pathway," whether it is associated with recovery, plasticity, or some other phenomenon, there would be no cortical or event-related potentials. What this suggests is that the disruption of neural impulses at the level of the midbrain can deprive the cortex of the necessary input needed to generate its potentials. It additionally suggests that there is no other major pathway from the brainstem to cortex other than that which courses through the inferior colliculi that can provide the synchronous input to the subcortical and cortical pathways needed to generate the middle and late potentials.

As with the MLR, it has been reported that late evoked potentials can be obtained even when the ABR is absent or severely compromised (Squires and Hecox, 1983). However, in these cases one must entertain the possibility that there may have been some plasticity factors involved (as discussed earlier) and that perhaps even with an absent ABR the inferior colliculus could have been sufficiently intact to conduct the neural impulses required for the generation of the late potentials. It is well known that the inferior colliculus is the main synaptic station of the brainstem pathway and that when it is severely damaged input to the next level will be drastically reduced (van Noort, 1969). It is reasonable to assume that since the inferior colliculus is larger than any of the more caudal auditory brainstem nuclei and

since most of the ascending fibers from the more caudal nuclei synapse at the inferior colliculus, damage to the inferior colliculus would have a greater effect on input to the cortex than damage to the lower brainstem structures (see Pickles, 1988 for review). Given this scenario, it is logical to assume that inferior colliculus damage of a severe degree would have greater influence on the propagation of mid and late potentials than the lack of an ABR. Hence, it is our belief that the mid and late potentials (and other auditory processing at cortical levels) are highly dependent on the integrity of the inferior colliculi and that if CANS function at this level had not improved in this patient over time that the mid and late potentials would have remained absent throughout the period that this individual was followed.

The lack of "normal" auditory evoked potentials at 10 months was more consistent with the patient's symptoms than were the results of basic pure-tone and speech testing. The patient still had problems hearing in noise and in other complex listening environments. It could be that the evoked potentials and listening in noisy backgrounds required a level of auditory processing sophistication not yet supported by a partially recovered neural substrate. These findings clearly support the importance of using electrophysiologic measures to document auditory difficulties in many patients with CANS involvement.

Intensity Discrimination

Intensity discrimination was measured at five and 11 weeks. At five weeks the right ear could not be tested due to poor threshold sensitivity and inconsistent responses at the frequencies tested (500, 1000, 2000 Hz). At 11 weeks, there was a rather marked improvement in the patient's performance at all frequencies, with the most significant change noted at 2000 Hz. In addition, a clear binaural advantage for the intensity discrimination task was observed. A similar binaural advantage was also noted during speech recognition testing in this patient, as well as in other centrally deaf subjects (Musiek et al, 1994). At 11 weeks, the patient's intensity discrimination, although noticeably improved, was not normal, and this may have been the key reason why the speech recognition scores remained poor at this time.

We believe there is a relationship among abnormal difference limens (DLs) for intensity and frequency and poor speech recognition scores. If the intensity (or frequency) discrimination is so poor that one cannot recognize changes in intensity (or spectral information) from phoneme to phoneme, then speech perception will suffer. Cranford and colleagues (1982) in an investigation of patients with temporal lobe lesions showed increased DLs for frequency in ears that also demonstrated reduced speech recognition scores. This was especially evident when the tone duration used in the DL test was relatively brief. We also know of at least one previous case of a patient with word deafness in which the DLs for frequency were found to be extremely large (Musiek et al, 1994). At this time it is not possible for us to indicate whether this is a common finding in patients with central deafness since few of the previous investigations reported in the literature have undertaken this type of assessment. With additional testing, a clear relationship between intensity and frequency discrimination abilities and speech recognition performance may be documented.

Rehabilitation

The rehabilitation program for this patient focused primarily on auditory training with some work on language use and the utilization of visual cues. It is difficult to know how much of a roll the therapy played in this patient's auditory improvement. Certainly much of the patient's progress was related to spontaneous recovery given the time period over which most of the progress was made. However, it is known that recovery can often be enhanced by stimulation and by presenting tasks to the central nervous system that challenge it (see Musiek and Berge, 1998, for review). It is therefore likely that the patient's improvement was positively affected by the therapy activities in which he participated. The patient's progress in therapy is reflected by the increased sophistication in the tasks from the beginning to the end of the formal sessions. At the beginning, the patient's identification and discrimination had to do with gross, highly different environmental sounds. At the end of therapy he was working on differentiating words, phonemes, and CVs that were highly similar in their phonetic features. There is

evidence that auditory training can improve auditory abilities related to discrimination and temporal processing (Recanzone et al, 1993; Kraus et al, 1995a, 1995b; Tallal et al, 1996), as well as other higher-level linguistic processes. Therefore, it seems likely that the therapy did make a contribution to this patient's recovery.

SUMMARY

There are a number of key observations from this study that may be of interest to the clinician and investigator. To our knowledge, this is the first report of a case of central deafness associated with damage to both inferior colliculi. The audiological testing of this patient demonstrated a wide range of auditory deficits associated with profound damage to both inferior colliculi. These findings were consistent, for the most part, with those noted for other cases with central deafness originating from other structures located more rostrally within the central auditory nervous system. The most notable difference was the observation of abnormalities of wave V of the ABR. Although such abnormalities would not be expected in cases with compromise of cortical or subcortical structures, these abnormalities would also not have been predicted for the present case based upon our current understanding of the generator sites for the ABR waves (Moeller, 1983). This finding suggests that at least in this case the inferior colliculus contributes to some degree to the generation of wave V of the ABR. Also of interest in this case was the observation of a hierarchical course of recovery with the patient demonstrating the ability to process simple acoustic stimuli and tasks before he could process auditory functions related to more complex acoustic stimuli and tasks. Unlike his performance on pure-tone, spondee threshold and speech recognition testing, his performance on other tests such as auditory evoked potentials and intensity discrimination did not recover to normal values at 10 months. This may indicate a disassociation in the physiologic bases of these different types of tests and/or that these types of tests have a different level of processing sophistication. Finally, this case adds to the growing body of evidence that central auditory involvement can affect pure-tone sensitivity.

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